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CLINICAL REPORT

Femoral parosteal osteosarcoma 18 years after its discovery: A case report

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KEYWORDS
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Summary Osteosarcomas are a heterogeneous group of tumors with diverse anatomical, clinical, and progressional characteristics. Parosteal osteosarcoma, or juxtacortical osteosarcoma, is a rare form of osteosarcoma that develops at the bone surface, but has a better prognosis than other conventional osteosarcomas. We report the observation of a 22-year-old female patient whose initial presentation was an enormous tumefaction of the knee that had been progressing for 10 years. The biopsy concluded in PO of the lower third of the femur. Staging was negative. The tumor had reached an enormous size and required amputation of the left lower extremity. A custom external prosthesis was manufactured to get her back to walking. Eight years after surgery, no local recurrence or metastasis has been detected. Parosteal osteosarcoma is a rare form of osteosarcoma with very slow progression (in spite of the particularly dramatic presentation in our observation), with an excellent prognosis and very rare metastasis.

Introduction

Parosteal osteosarcoma (PO), or juxtacortical osteosarcoma (JCO), is a rare anatomical and clinical variant of osteosarcoma, accounting for 4% of cases [1,2]. It progresses slowly and has a good prognosis compared to conventional osteosarcoma. The authors report an original observation that illustrates the low aggression of this tumor and discuss its anatomical and clinical features.

Observation

We report the case of a female patient, born in 1978, consulted in 1995 at the age of 17 years for a painless mass in the right knee that had appeared at the age of 12 years (1990) and progressively increased in volume. The physi-

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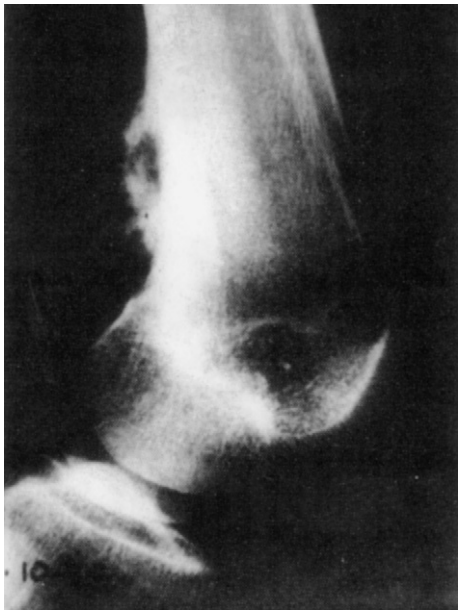

Figure 1 The first X-ray of the knee (at the age of 17 years): mass of the lower one third of the femur, dense, relatively well limited, attached to the metaphyseal cortex by a wide base.

Figure 2 Enormous tumor mass measuring 40 cm at its widest point, limiting all knee movement.

cal examination revealed a 2 cm mass at its largest above the right popliteal fossa with knee flexion moderately limited. The X-ray showed a mass in the lower third of the femur that was dense, relatively well limited, attached to the metaphyseal cortex by a wide base (Fig. 1). The biopsy showed moderately cellular malignant tumor proliferation, with slightly atypical and rarely mitotic fusiform cells surrounded by osteoid trabeculae, providing a diagnosis of JCO. Conservative treatment (wide resection of the tumor with knee prosthesis) preceded by a course of first-line chemotherapy was planned at this stage. However, the patient only received two cycles and then was lost to follow-up. Two years later, in 1997, with continued progression and no response to the chemotherapy, amputation was proposed but refused by the patient. She returned 3 years later (2000), at the age of 22, with a considerable increase in tumor volume. The physical examination showed a tumor mass measuring 40 cm at its widest point, limiting all knee movement (Fig. 2). The plain X-ray showed voluminous lytic and condensing bone tumor, occupying the lower third of the femur extending to the middle third with invasion of the soft tissues (Fig. 3).

Magnetic resonance imaging (MRI) showed a voluminous heterogeneous tumor, measuring approximately 35 cm $\times$ 25 cm, invading the soft tissues, the knee joint, the upper third of the tibia and fibula, as well as the medullary cavity, encompassing the vessels. This tumor was slightly hypointense compared to the muscle and included areas of hypointense signal on T1-weighted sequences, with no enhancement after gadolinium injection. It was located approximately 10 cm from the center of the femoral head, with no skip metastases (Fig. 4).

The staging bone scintigraphy and thoracoabdominal CT showed no secondary extension. The patient was treated by amputation at the hip, performed on 20 May 2000, 10 years after the first symptoms.

The anatomopathological examination of the specimen showed a 42 cm $\times$ 38 cm tumor of the knee and thigh. It was located 7 cm from the limit of surgical resection. The cut specimen presented a lobulated, whitish aspect with no notable cartilaginous zones. The skeletal muscles, the knee joint, and the upper end of the tibia and fibula were invaded by the tumor.

The histological analysis showed a malignant mesenchymatous proliferation, moderately cellular, made up of long, linear and eosinophilic material, sometimes calcified with no

Figure 3 X-ray 5 years later (age, 22 years): voluminous lytic and condensing bony tumor occupying the lower third of the femur extending to the middle third and invading the soft tissues.

Figure 4 MRI: heterogeneous tumor, slightly hypointense signal compared to muscle, with T1-weighted hyposignal areas, with no enhancement after gadolinium injection. The tumor invaded the soft tissues of the knee and the medullary cavity with no skip metastases.

osteoblastic cells in the periphery, corresponding to osteoid (Fig. 5). The tumor cells were spindle-shaped, with little eosinophil cytoplasm and a long or ovoid, hyperchromatic, and moderately atypical nucleus. Mitoses were rare (Fig. 6). There were no areas of dedifferentiation. We diagnosed juxtacortical (parosteal) osteosarcoma with no dedifferentiation areas, with healthy excision margins.

A walking prosthesis was prescribed that allowed the patient to walk autonomously with no discomfort or assistance. She was followed-up in outpatient consultation until February 2008, with a last follow-up X-ray (of the thorax) and scintigraphy (bone scintigraphy) showing neither recurrence nor metastasis 18 years after the first symptoms.

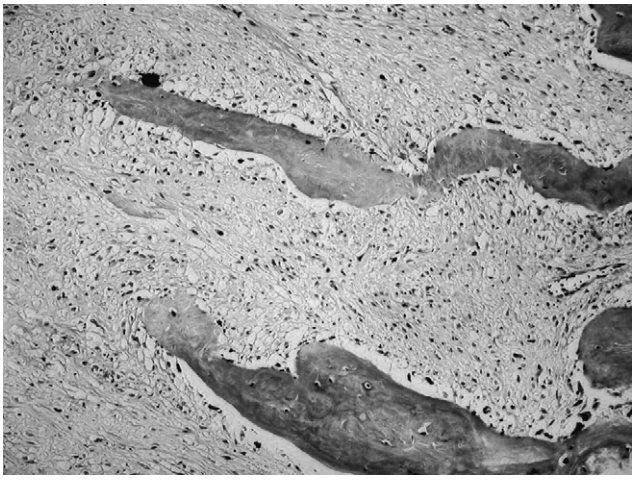

Figure 5 Histology of parosteal osteosarcoma: moderately cellular proliferation with long osteoid trabeculae with no osteoblastic areas (magnification $\times$ 250).

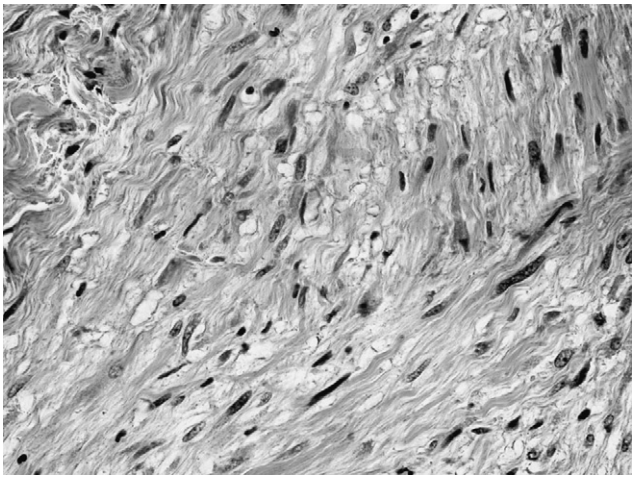

Figure 6 Greater magnification ($\times$ 400). Tumor cells are spindle-shaped with a long or ovoid nucleus, hyperchromatic, and moderately atypical.

Discussion

PO, or JCO, is a rare disease described for the first time by Gerschickter and Copeland in 1951 [3]. It is the most frequent form of bone surface osteosarcoma [1,2]. It is a well-differentiated, low-grade disease.

PO onset is on average 10 years later than conventional osteosarcoma [4,5]. Age at onset is between 15 and 40 years, with a mean of 28 years [2,6,7]. There seems to be a female predominance, with a sex ratio of 1:3. Our patient was a 12-year-old girl at the time of diagnosis. PO usually presents with a globular, juxta-articular mass that is fairly well limited, hard in consistency, slow-growing, generally painless, but it can cause functional discomfort, limiting joint movement in one third of cases [6,7]. The painless quality of the disease explains the delay in diagnosis. In our observation, the tumor evolved progressively and slowly for 10 years, reaching an impressive size.

The most frequent seat is the distal end of the femur, where the tumor arises typically on the posterior side. This location is followed by the proximal end of the tibia, then rarely by the proximal end of the femur, humerus, and radius [1,5]. PO cases reaching the carpal or tarsal bones and the flat bones have been described [7].

The radiology aspect is typical. PO presents as a radiopaque, metaphyseal mass developing on the external side of the cortex, extending toward the soft tissues. This mass is dense and homogeneous, with polycyclic or archiform contours. At an early stage, there is a radiotransparent groove that separates the tumor from the bone cortex except at the base of the implantation. When it is present, this sign has a high diagnostic value. It disappears in voluminous tumors. According to Edeiken-Monroe et al., this clear ring corresponds to the periosteum between the tumor and the bone cortex [7,8]. Periosteal reaction, frequent in conventional osteosarcoma, is absent in PO. In our case, the first X-ray showed a dense, quite well-limited mass, attached to the metaphyseal cortex by a wide base. The clear space between the tumor and the cortex is not very sharp on the X-ray image.

CT provides a better image of the radiotransparent groove, clarifies the tumor's implantation base, and demonstrates any intramedullary extension or satellite lesions. MRI is the choice examination to detect intramedullary invasion and elucidate the tumor's position in relation to vascular and nerve structures [7,9]. In our case, MRI demonstrated a voluminous heterogeneous tumor invading the soft tissues, the knee joint, the upper third of the tibia and fibula, and the medullary cavity, including the vessels.

The macroscopic aspect is typical. The tumor presents as a voluminous, exophytic, ossified mass, with well-limited contours, attached to the bone by a wide base, pushing the adjacent structures back. The cut specimen has a heterogeneous aspect, showing some fibrous zones and some cartilaginous zones. Intramedullary extension is present in 25% of cases and should be meticulously searched for in multiple samples. The existence of areas of soft and fleshy consistency suggest dedifferentiation [1]. In our case, these zones of soft and fleshy consistency were not found.

Histologically, the tumor comprises parallel osteoid trabeculae with or without a peripheral osteoblastic area. These trabeculae are separated by cellular proliferation, made up of spindle-shaped cells presenting minimal atypia and a low level of mitotic activity [1,2].

In 50% of cases, the tumor presents areas of cartilaginous differentiation in the form of nodules or a cap in the surface [1]. In the case reported here in, we found no cartilaginous nodules. Three histological grades are distinguished: grades I and II, corresponding to the conventional form, with a low level of malignity. Grade III, corresponding to the dedifferentiated form with manifest atypia and mitoses, has a high level of malignity.

Dedifferentiation is observed in 15% of cases. It is present at diagnosis or most often at recurrence. This dedifferentiation can manifest as osteosarcoma, fibrosarcoma, or malignant histiocytoma [1]. In the case reported herein, no areas of dedifferentiation were found and the tumor was low grade (conventional PO).

The differential diagnosis is made essentially with benign lesions such as osteoma, osteochondroma, ossifying myositis, and periosteum desmoid. Clinical and radiological data contribute enormously to the diagnosis. Histologically, these lesions do not develop in a sarcomatous stroma and the cells are regular with no atypia or hyperchromatism. The differential diagnosis is also made with low-grade periosteal osteosarcoma and intramedullary osteosarcoma. The radiological presentation is different and there is no clear space between the tumor and the cortex. From a macroscopic viewpoint, there is a topographical difference between the tumor and the bone structures [10].

The recommended treatment is surgery consisting of wide resection of the tumor in a single piece. According to Enneking, resection can be intracapsular, marginal, wide, or radical. Amputation is reserved for cases in which wide resection cannot be performed, as in our patient, or when the tumor is dedifferentiated or recurrent. Adjuvant chemotherapy can be effective in the dedifferentiated forms [11]. Our patient received a course of primary chemotherapy but did not respond. The prognosis of PO is good if the patient is properly treated. Overall survival at 5 years is of the order of 91% [1]. The risk of local recurrence depends on the quality of the excision. It is 50% if the resection is marginal, passing in a peritumoral reactive zone [12,13]. If resection is total, recurrence is exceptional. Rare cases of tumor recurrence after a long period have been reported: 15 years in Okada and Swee's series and 20 years in the case reported by Lau et al. [4,14]. Metastases are seen in 38% of cases. They are generally associated with dedifferentiated or recurrent forms or those with medullary invasion. The most frequent site is the lungs, rarely the abdomen and central nervous system, and exceptionally the myocardium [14]. In our patient, during a slow progression over 10 years before intervention, followed-up for 8 years after amputation, no recurrence or metastasis has been observed.

Conclusion

PO is a low-grade malignant bone tumor characterized by its insidious progress and good prognosis. It rarely leads to metastasis. Its treatment is essentially surgical. Areas of dedifferentiation are the main factor indicating poor prognosis. Our observation illustrates that this tumor is not aggressive, with no metastasis after 18 years of progression.

References

[1] Unni KK, Knutila S. Parosteal osteosarcoma. In: World Health Organization Classification of Tumours. Tumours of soft tissue and bone. 2002. p. 279—81.
[2] Fechner RE, Mills SE. Parosteal osteosarcoma. Atlas of tumour pathology. In: Tumours of the Bones and Joints. 1992. p. 65—70.
[3] Gerschickter CF, Copeland MM. Parosteal osteosarcoma: a new entity. Ann Surg 1951;133:790—807.
[4] Okada K, Swee RG. High-grade surface osteosarcoma: clinicopathologic study of 46 cases. Cancer 1999;85:1044—54.
[5] Drinkuth S, Segmüller H, Furrer M, Wartburg UV. Parosteal osteosarcoma of the distal ulna. A rare tumour at a rare location: a case report. Chirurgie de la main 2003;22:104—8.
[6] Babinet A, Tomeno B, Forest M. Ostéosarcome de basse malignité. EMC, Appareil locomoteur, Elsevier-Paris, 14-708, 2001, 6p.
[7] Edeiken-Monroe B, Edeiken J, Jacobson HG. Osteosarcoma. Semin Roentgenol 1989;24:153—73.
[8] Temple HT, O'Keefe RJ, Katapurum S, Mankin HJ. Clinical outcome of 38 patients with juxtacortical osteosarcoma. Clin Orthop 2000:208—11.
[9] Okada K, Frassica FJ, Sim FH, et al. Parosteal osteosarcoma: a clinicopathological study. J Bone Joint Surg (Am) 1994;76:366—78.
[10] Enneking WF. Musculoskeletal tumours surgery, 2. New York: Churchill Livingstone; 1983.
[11] Steenbrugge F, Poffyn B, Uyttendaele D, Verdonk R, Verstraete K. Parosteal osteosarcoma of the humerus. Acta Orthopaedica Belgica 2001;67:387—94.
[12] Bertoni F, Bacchini P, Staals EL, Davidovitz P. Dedifferentiated parosteal osteosarcoma: the experience of the Rizzoli institute. Cancer 2005;103:2373—82.
[13] Lau TW, Shek TWH, Wong LLS. Local recurrence of parosteal osteosarcoma adjacent to a prosthesis after 20 years: a case report. J Orthop Surg 2004;12:263—6.
[14] Santos-Machado TM, Mendes Oliveira CR, Croci AT, Fernandez A, Abadi DM, Baptista AM, et al. Parosteal osteosarcoma with myocardial metastasis 13 years after follow-up. Rev Hosp Clin Fac Med S Paulo 2003;58:113—8.